

Neonatal ovarian cysts: therapeutic dilemma

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SUMMARY Seven cases of neonatal ovarian cysts that presented over the past seven years were studied. Complications included torsion and rupture and usually occurred in cysts more than 5 cm in diameter. Surgical removal, either oophorectomy or cystectomy, was the treatment of choice. Because even cystectomy results in loss of normal ovarian tissue, and because spontaneous regression of cysts less than 5 cm in diameter can occur, a more conservative approach is now proposed. Regular ultrasonography alone is recommended if the cysts are less than 5 cm in diameter, and aspiration of the cysts followed by regular ultrasonographs if the cysts are more than 5 cm in diameter. Operation should be reserved for recurrent cysts or for those with complications. Cysts diagnosed antenatally may be aspirated in utero if there are signs of thoracic compression.

Neonatal ovarian cysts are being diagnosed more often now that routine ultrasonography is carried out antenatally and postnatally. A truly cystic abdominal mass in a baby girl is most likely to be an ovarian cyst, although duplication cyst or mesenteric cyst should be considered in the differential diagnosis.

There is controversy about the best treatment for these cysts, opinions ranging from oophorectomy to follow up by ultrasonography alone. We propose a regimen the main objective of which is to avoid unnecessary operation especially in those patients with bilateral cysts.

Case reports

CASE 1

The infant was born prematurely by emergency caesarean section at 33 weeks' gestation weighing 2700 g. The mother had insulin dependent diabetes and had been treated for thyrotoxicosis by thyroidectomy six years previously. She had also had one spontaneous abortion and two stillbirths.

A routine antenatal ultrasound scan at 32 weeks showed a cystic mass in the baby's abdomen. This was confirmed when she was 2 weeks old as a cystic abdominal mass measuring 4×3 cm. At laparotomy on the following day a cyst arising from the right ovary was enucleated. The uterus and left ovary were normal. On histological examination the cyst was found to be a simple follicular cyst with evidence of luteinisation. The patient made an uneventful recovery.

CASE 2

The infant was born by normal spontaneous vaginal delivery at 35 weeks' gestation weighing 3200 g. The mother had had two spontaneous abortions and two pregnancies terminated; she was also a heroin addict. Nine days before delivery two cystic central abdominal masses that were separate from the kidneys were noted on ultrasound scan (fig 1a). A scan performed when she was one day old (fig 1b) confirmed the presence of cystic structures measuring 8×4 cm and 5×4 cm diameter, respectively. At operation bilateral ovarian cysts were found and bilateral cystectomies performed. Both cysts were unilocular and contained clear yellow fluid. On histological examination the cyst walls were found to be thin and contained some ovarian tissue with a few follicular cysts. The patient made a satisfactory recovery and was discharged home three weeks after the operation.

CASE 3

The infant was born by normal vaginal delivery at full term weighing 3300 g. The mother was well. An ultrasound examination was performed at 38 weeks' gestation for a possible abruptio placentae and showed a cystic lesion 6 cm in diameter in the right side of the baby's abdomen. This was confirmed by a scan performed when she was 1 day old, and the lesion contained a few fine septa. At laparotomy on day 8 a mobile, right sided ovarian cyst was found, which had twisted twice at the junction of the right fallopian tube and the uterus. A right salpingo-oophorectomy was performed. The left ovary was

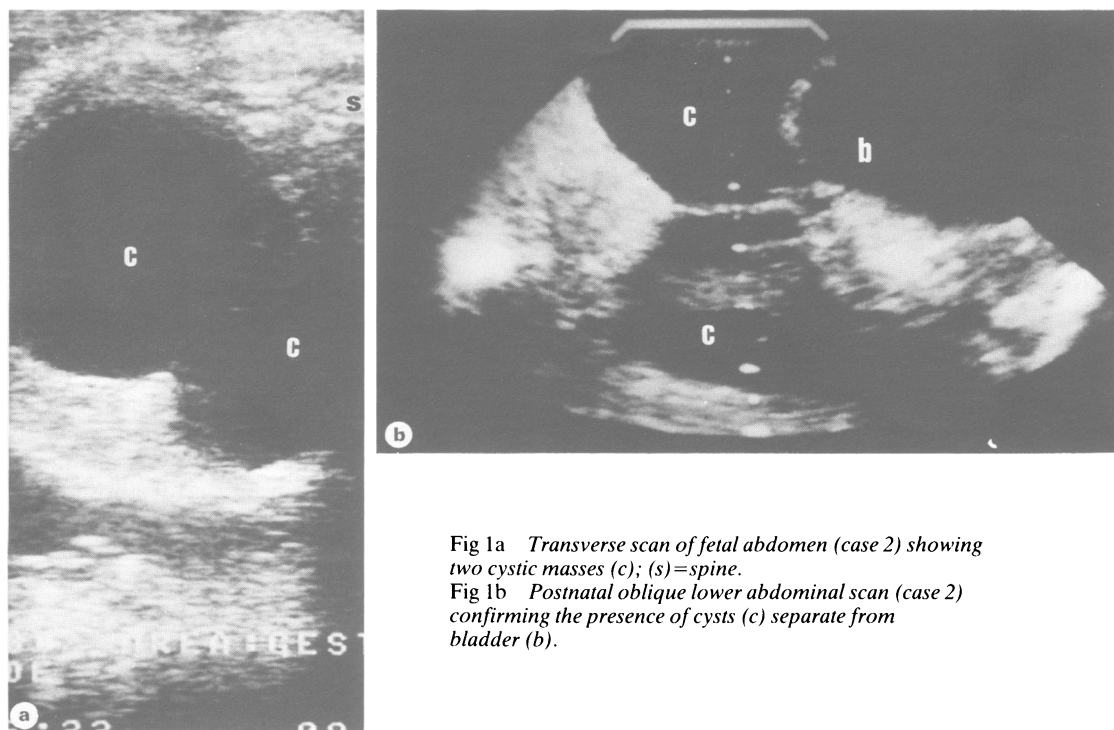


Fig 1a Transverse scan of fetal abdomen (case 2) showing two cystic masses (c); (s)=spine.

Fig 1b Postnatal oblique lower abdominal scan (case 2) confirming the presence of cysts (c) separate from bladder (b).

normal and contained a few small follicular cysts. An infarcted cyst was confirmed on histological examination. The cyst was thickened (1 cm) in places and contained dark brown fluid. The patient was discharged on the sixth day postoperatively after an uneventful recovery.

CASE 4

The infant was born by spontaneous vaginal delivery at 27 weeks' gestation weighing 2000 g. She had many problems in the immediate neonatal period including respiratory distress syndrome that required ventilation, a grade II intraventricular haemorrhage on the left, jaundice, and haemolytic anaemia that required two transfusions. No cause was found for the anaemia but during investigation an ultrasound examination was performed to exclude polysplenia. The spleen was normal but a cystic lesion in the abdomen was found. A laparotomy was performed at 18 weeks at which a twisted cyst of the right ovary 5 cm in diameter was found together with a cyst of the left ovary 2.5 cm in diameter. Right salpingo-oophorectomy and drainage of the left cyst with biopsy were carried out.

On histological examination the specimen from the left cyst showed normal ovarian tissue whereas the right cyst was totally infarcted with areas of calcification within it and only a simple flattened epithelial lining; it contained brown necrotic material. The patient was discharged home 13 days postoperatively.

CASE 5

The infant was born by normal vaginal delivery at full term weighing 3800 g after an uneventful pregnancy. The mother was well. At the age of 5 months the baby was admitted to hospital with abdominal distension and a three month history of intermittent abdominal swelling that had caused dyspnoea at night. Ultrasound examination showed a unilocular cyst 10 cm in diameter in the centre of the abdomen, separate from the kidneys and the bladder. At laparotomy a large left sided ovarian cyst was found that had twisted two and a half times round the left fallopian tube. A cystectomy was performed, and an ovarian remnant and the left fallopian tube were left behind. The right adnexa was normal. On histological examination the cyst contained dysplastic ovarian parenchyma with a few

oogonia and an occasional primary graafian follicle. The patient was discharged home six days post-operatively.

CASE 6

The infant was born by normal vaginal delivery at full term weighing 3300 g. The mother was well. A routine antenatal ultrasound scan at 32 weeks' gestation showed a cystic mass 4×3 cm in diameter in the left side of the abdomen separate from the kidneys. This was confirmed when she was 5 weeks old, but this time the lesion lay on the right side of the abdomen. The mass was not palpable. By 25 weeks of age the mass had become palpable and repeat ultrasound examination confirmed an increase in its size to 6×5 cm. A large cyst of the left ovary was removed at laparotomy. The right ovary was normal. She also had a malrotation of the bowel and this was corrected. On histological examination the ovarian mass was found to be a simple follicular cyst. The patient made an uneventful recovery.

CASE 7

The infant was born by normal vaginal delivery at full term weighing 3100 g. The mother was well, but the pregnancy was complicated by hydramnios. A routine antenatal ultrasound scan at 34 weeks' gestation showed a cystic mass 7×5 cm in diameter in the abdomen (fig 2). This was confirmed when she was 1 day old, the mass having decreased in size to 6×4 cm. The patient was otherwise normal, in particular there was no evidence of gut atresia to account for the hydramnios. A scan two weeks later suggested that the mass was multilocular. At laparotomy bilateral ovarian cysts were found, the left being about twice the size of the right. The right cyst was chronically twisted and a right salpingo-oophorectomy was performed, together with a left cystectomy. On histological examination the right cyst was found to be multilocular and follicular, and the left unilocular and follicular. There were no postoperative complications and the patient was discharged home three days later.

Discussion

Before the introduction of ultrasonography, ovarian cysts in neonates were thought to be rare and could only be diagnosed postnatally.¹⁻³ Only 71 cases were reported before 1976.⁴ Cysts were only discovered if they were palpable or became symptomatic, but, with ultrasonography asymptomatic ovarian cysts could be diagnosed.^{5,6} Ovarian cysts can now be diagnosed antenatally, and 11 such cases had been reported up to 1987.^{7,8} The pathogenesis of neonatal ovarian cysts is unknown. Examination of

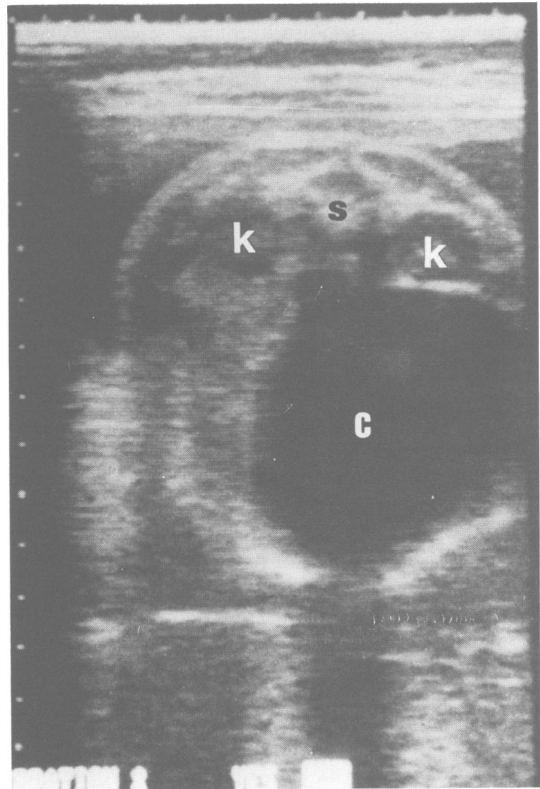


Fig 2 Transverse section of fetal abdomen (case 7). Large abdominal cyst (c) is shown separate from kidneys (k) and spine (s).

the ovaries of 121 children at necropsy showed that there was more pronounced luteinisation of the theca interna in the neonatal ovaries studied when compared with the older age groups⁹; this was subsequently confirmed by a study of normal neonatal and infantile ovaries.¹⁰ Another series of 332 necropsies of stillbirths and neonatal deaths showed that 34% had small follicular cysts within 28 days of birth.¹¹ It is almost certain, therefore, that the stimulus for the formation of neonatal ovarian cysts is chorionic gonadotrophin that stimulates the fetal ovary during pregnancy. Luteinising cysts are occurring increasingly more often in babies whose mothers were diabetic (case 1) or had toxæmia, or maternal isoimmunisation, because these conditions are all associated with raised concentrations of human chorionic gonadotrophin.^{12,13} There is also an increasing incidence in premature infants, probably because of their greater sensitivity to human chorionic gonadotrophin (cases 1, 2, and 4).¹⁴ On

histological examination the cysts are follicular or luteinising cysts. Because human chorionic gonadotrophin is important in the pathogenesis of neonatal ovarian cysts, it is logical to expect the cysts to regress spontaneously in the neonatal period as the hormone concentrations fall and the stimulus for growth disappears. This often seems to happen unless some complication supervenes.^{8,12} All the reported cases of spontaneous resolution have occurred within four months of birth.

Both sides seem to be affected with equal frequency, also a finding in our series. Bilateral cysts were said to be rare, only three cases having been described by 1974,¹² though the incidence has increased since the introduction of ultrasonography; three of our cases had bilateral cysts.

Malignant change in neonatal ovarian cysts that are simple fluid filled structures is extremely rare, and is usually seen only in more complex lesions.¹ Other complications, however, are common. Most commonly reported complications of untreated ovarian cysts are torsion of the pedicle (cases 3, 4, 5, and 7), haemorrhage into the cyst or abdominal cavity, and rupture of the cyst.^{1,3,5,6,15}

The cysts may be of any size, the largest reported being over 20 cm in diameter. The larger cysts are associated with other complications, including intestinal obstruction with one reported case of caecal perforation in an infant,¹⁶ and hydramnios. One of our cases was associated with hydramnios (case 7) and another probably had intermittent intestinal obstruction (case 5). Hydramnios has been reported in 5 to 10% of pregnancies in which a fetal ovarian cyst was subsequently diagnosed.^{12,17,18,19} This is probably caused by pressure of the mass on the small bowel together with interference with the swallowing mechanism of the fetus, which reduces ingestion and absorption of amniotic fluid.

There is a risk of pulmonary hypoplasia developing in fetuses with large cysts.⁷ Pulmonary hypoplasia is associated with other lesions which compress and reduce the intrathoracic space (for example, large abdominal fluid collections and masses, and diaphragmatic hernias and eventration). Though there have been no definite cases reported of pulmonary hypoplasia developing secondary to an ovarian cyst, the risk exists if the cyst is large enough to cause thoracic compression as with any other large abdominal mass.

Nowadays the diagnosis of ovarian cysts is by ultrasound scan and may be made antenatally or postnatally. Mesenteric and duplication cysts may, however, be ultrasonically indistinguishable from ovarian cysts.⁶ Most neonatal ovarian cysts are asymptomatic and found incidentally. Some of the

cysts are extremely mobile and appear as a 'wandering' tumour (as seen in cases 3 and 6.)⁵

The treatment of these ovarian cysts is controversial. Until recently the recommended treatment was surgical. If the cyst was unilateral or if there had been torsion, oophorectomy was performed. The rationale for this when a simple unilateral cyst was present was the risk of possible torsion, or the replacement of the entire ovary by cyst making cystectomy impossible, or the inability to distinguish cyst from remaining normal but distorted ovarian tissue on histological examination.^{3,4,6} More recently simple cystectomy was recommended but whether any function remained in the ovary is unknown.⁶ The cystectomy specimens in our series all contained normal ovarian tissue, thus supporting the above findings. This is not of any great importance if there is a normal ovary on the other side.

Problems arise, however, when the cysts are bilateral. Oophorectomy on one side and cystectomy on the other have been done, as have bilateral cystectomies. These operations carry a high risk of removing all normal ovarian tissue (despite attempts to preserve some of each ovary), thereby rendering the patient sterile.

All the cases in our series had either cystectomy or salpingo-oophorectomy. We therefore decided to see whether alternative and more conservative treatment had been reported, and, indeed surgical conservatism had been advocated for some time and the advent of cyst aspiration was predicted in 1972.³

In a recent paper, Nussbaum *et al* described three cases in which cysts had resolved spontaneously within four months of discovery.⁸ All were less than 5 cm in diameter and were followed up by serial ultrasound scans. Though there is a potential risk of torsion, only two cases have been reported in cysts of less than 5 cm in diameter.^{5,6} In both cases the torsion was detected on the ultrasound scan because the previously echo free cyst developed fluid and debris levels within it. Both cases underwent operations, though it is not clear whether there is a risk in leaving a twisted, infarcted cyst untreated. Rupture of the cyst is rare and has only been reported in larger lesions (10 to 12 cm in diameter); it would not therefore seem to be a risk factor in lesions less than 5 cm in diameter.¹⁵

Lesions more than 5 cm in diameter present a greater problem as there is an increased risk of complications if they are left untreated. In view of the natural tendency of these cysts to regress spontaneously we suggest that cyst puncture should be performed, and the patient followed up with serial ultrasound scans. Recurrence of the cyst could be treated by repeated aspiration, surgical removal

being reserved for the few intractable or complicated cases. There have been two recent case reports of intrauterine ovarian cyst aspiration in which there was thought to be an appreciable risk of pulmonary hypoplasia secondary to a pronounced degree of thoracic compression.⁷ The most important risk associated with cyst puncture is cyst rupture leading to peritonitis. This is, however, uncommon.²⁰

Oestradiol concentrations should be measured in aspirated fluid to confirm its ovarian origin,²¹ though high levels of oestrogen are found in the normal neonatal circulation.²² This oestrogen, the exact structure of which is unknown, is probably manufactured and secreted by the fetal adrenal glands and may interfere with the oestradiol estimation by some of the direct assay kits used in some laboratories. Spuriously high levels of oestradiol may, therefore, be detected in the aspirate if the sample is contaminated with blood. This should be borne in mind if

the laboratory uses these kits.²³ A high oestradiol concentration in clear aspirate from a cyst indicates with certainty that it is of ovarian origin.

A conservation approach to this problem should therefore be attempted before embarking on surgical treatment. The advantages of this are firstly, that the patient may require no treatment if the cyst regresses spontaneously on serial ultrasound scans. Secondly, if cyst aspiration is effective then the patient is spared an operation, and thirdly—perhaps most importantly—if the cysts are bilateral the conservative approach prevents the possible removal of all normal ovarian tissue. We therefore propose therapeutic regimens for the treatment of uncomplicated neonatal ovarian cysts (tables 1 and 2). Complicated cysts should be treated with as conservative an operation as possible.

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Table 1 *Regimen for antenatal diagnosis of uncomplicated neonatal ovarian cysts*

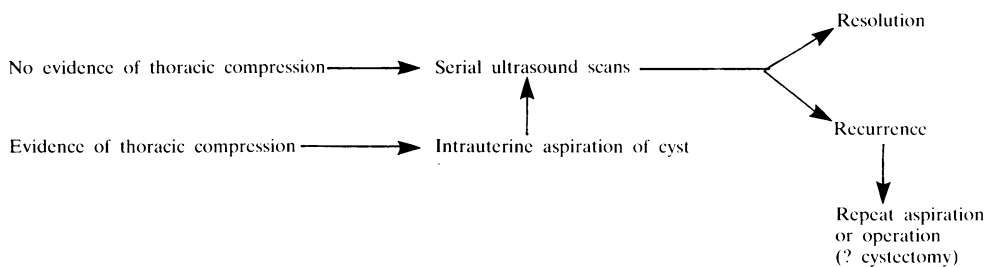
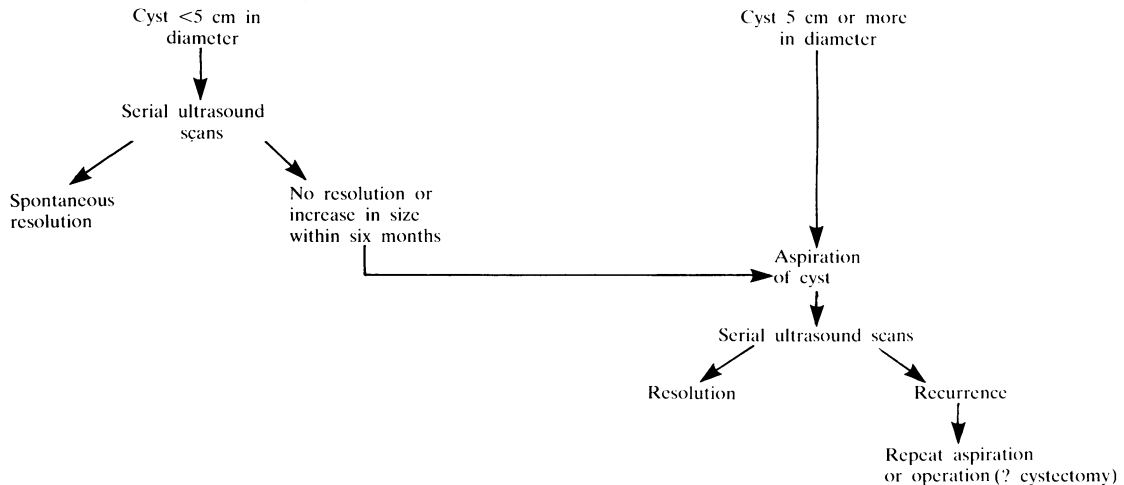


Table 2 *Regimen for postnatal diagnosis of uncomplicated neonatal ovarian cysts*



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